BRIEF COMMUNICATION

Endoscopic Electrocauterization for Congenital Pyriform Sinus Fistula Treatment in Paediatrics. Case Series

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Abstract Pyriform sinus fistulas are rare anomalies of the branchial arches. Most of them are located on the left side. They extend from the apex of the pyriform sinus of the hypopharynx to the thyroid gland or adjacent tissues.

The diagnosis is suspected in the presence of acute suppurative thyroiditis or recurrent cervical abscesses, and is confirmed by endoscopic visualisation of the fistula hole. The traditional treatment consists of excision of the fistulous tract, with or without thyroid lobectomy, by cervical approach. However, less invasive alternatives that obliterate the path of the fistula have been developed, such as endoscopic electrocautery.

We describe our experience with 7 patients with this condition, who were treated with endoscopic cauterization using radiofrequency electrocautery, and we evaluate the effectiveness and safety of the treatment performed.

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Introduction

Pyriform sinus fistula (PSF) is a rare congenital anomaly caused by incomplete obliteration of the third or fourth pharyngeal bursa during the seventh week of gestation.\(^1\)\(^2\) A more recent theory about the origin of the malformation suggests that it derives from the thymopharyngeal duct.\(^3\)\(^4\)\(^5\)

Clinically this causes suppurating acute thyroiditis and recurring lateral cervical abscesses.\(^6\) In the newly born it manifests as a compressive cervical mass that may hinder breathing and swallowing.\(^7\) Diagnosis is confirmed by direct laryngoscopy that makes it possible to visualise the fistula orifice in the pyriform fossa.\(^4\)\(^6\)\(^8\) An oesophagogram is useful to show the trajectory of the fistula, although it may give rise to false negatives.

Classically this condition is managed by treating the acute exacerbation with systemic antibiotics and by draining the abscess, followed by surgical resection of the fistula trajectory using a cervical approach.\(^1\)\(^7\) With the aim of reducing the morbidity of open surgery, different endoscopic techniques have been developed during the past 18 years to obliterate the fistula orifice. Monopolar diathermy cauterisation is the technique that is used most often.\(^7\)\(^9\)

Methods

7 patients with congenital PSF were examined retrospectively. They were treated endoscopically by the Respiratory Endoscopy Department over an 11 year period (September 2004–September 2015).

The variables analysed were patient sex, fistula location, age at the start of the symptoms and at the time of diagnosis, clinical manifestations, the diagnostic techniques used, the endoscopic treatment used, complications, the duration of follow-up and evolution.

The larynx and hypopharynx were examined endoscopically with a flexible fibrescope (3.5 mm) and local anaesthesia in the surgery, and with a rigid endoscope (5 mm) under general anaesthetic in the operating theatre, with the possibility of electrocauterisation if the diagnosis was confirmed. Images were recorded digitally.

Treatment consisted of the obliteration of the fistula orifice using radio-frequency electric scalpel cauterisation. Under general anaesthetic and with the patient in supine decubitus, cervical hyperextension and tracheal intubation, both pyriform sinuses are visualised using a laryngoscope and rigid 0° 5 mm optics. The laryngoscope is suspended to expose the pyriform fossa. Once the orifice of the fistula has been confirmed, a sheathed metal guide is inserted through it. The trajectory is obliterated using diathermy (Kairos Miniconp electrosurgical unit\(^a\)), from distal to proximal, at an intensity of 15 W until the mucus is seen to take on a white colouration. Antibiotic prophylaxis is administered (a 50 mg/kg/dose of ampicillin-sulbactam) intraoperatively.

Hospital discharge was permitted after 6–8 h observation. A normal oral diet was prescribed together with analgesics according to need (ibuprofen 10 mg/kg/dose).

The results were measured while taking into account the absence of clinical recurrence, the closure of the fistula orifice and the presence of complications.

All the patients were systematically subjected to a flexible fibrescopy with local anaesthesia one month after the endoscopic electrocauterisation, with clinical check-ups every 3 months during the first year and annual check-ups afterwards.

Results

There were 4 male patients (57.1%) and 3 female patients (42.9%) with PSF, aged from 11 months to 13 years old at
the commencement of symptoms. The form of presentation in 4 cases was a recurring cervical abscess and episodes of thyroiditis in 3. One patient, with exteriorisation of the fistula in the neck (secondary to the incision and drainage of the cervical abscess), presented leakage of liquids through the orifice in the skin while orally ingesting the same (Fig. 1A). Another patient presented purulent expectoration associated with the cervical infection.

In 6 cases (85.7%) the left side was involved, and the fistula was located in the right pyriform sinus in only one case (Fig. 2). The patient characteristics are shown in Table 1.

All the patients were diagnosed using a cervical ultrasound scan, and in some cases the study was completed using computerised tomographic imaging (CT) with contrast. Ultrasound scan and CT were useful in the evaluation of the inflamed mass and/or cervical abscess, although they did not identify the trajectory of the fistula.

A barium oesophagogram was performed in 3 patients 3 weeks after the resolution of the acute inflammatory phase.

<table>
<thead>
<tr>
<th>P</th>
<th>Sex</th>
<th>Age at start of symptoms</th>
<th>Age at diagnosis</th>
<th>Presentation symptoms</th>
<th>Fistula location</th>
<th>Relapse</th>
<th>Follow-up time</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>5 yr.</td>
<td>6 yr.</td>
<td>Thyroiditis × 2</td>
<td>Left</td>
<td>Yes</td>
<td>3 yr.</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>3 yr. 10 m</td>
<td>4 yr. 7 m</td>
<td>Cervical abscess</td>
<td>Left</td>
<td>No</td>
<td>9 m</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>4 yr.</td>
<td>4 yr. 6 m</td>
<td>Thyroiditis × 2</td>
<td>Right</td>
<td>No</td>
<td>9 m</td>
</tr>
<tr>
<td>4</td>
<td>F</td>
<td>11 m</td>
<td>2 yr. 10 m</td>
<td>Cervical abscess × 2</td>
<td>Left</td>
<td>No</td>
<td>6 m</td>
</tr>
<tr>
<td>5</td>
<td>M</td>
<td>9 yr.</td>
<td>10 yr.</td>
<td>Cervical abscess × 2</td>
<td>Left</td>
<td>No</td>
<td>2 yr.</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>1 yr. 10 m</td>
<td>2 yr.</td>
<td>Purulent expectoration</td>
<td>Left</td>
<td>No</td>
<td>6 m</td>
</tr>
<tr>
<td>7</td>
<td>F</td>
<td>13 yr.</td>
<td>17 yr.</td>
<td>Thyroiditis</td>
<td>Left</td>
<td>No</td>
<td>6 m</td>
</tr>
</tbody>
</table>

yr.: years; F: female; M: male; m: months; P: patients.
Only one patient presented leakage of contrast at the level of the pyriform sinus during transit of the oesophagus.

Rhinofibrolaryngoscopy with local anaesthesia was performed in all 7 patients, visualising the fistula orifice in 6 cases (Fig. 1B–D). In the remaining patient the fistula was not seen, although its presence was suspected as pyriform fossa asymmetry was observed. Direct laryngoscopy with rigid instrumentation under general anaesthesia confirmed the diagnosis of PSF in all of the cases.

The average age at the moment of diagnosis was 6.7 years old. The interval between the first signs and/or clinical symptoms and diagnosis varied from 2 months to 4 years.

Treatment of the initial acute phase consisted of broad spectrum systemic antibiotics, associated in 3 cases with drainage of the cervical abscess.

Once the acute phase had been resolved all of the patients were treated using endoscopic electrocauterisation (Fig. 3). In the patient with cervical exteriorisation of the fistula the orifice in the skin was cauterised with silver nitrate during the same session of anaesthesia.

No complications arose in association with the endoscopic procedure.

None of the fiberlaryngoscopies with local anaesthesia performed one month after the endoscopic procedure showed relapse of the lesion.

The average follow-up of the patients in outpatient facilities after the definitive treatment lasted for 1.14 years, and the parents were granted free access to return to the hospital if there were any problem. No relapse or recurrence of the symptoms were recorded during this time in 6 cases, which were resolved by a single session of electrocauterisation. Only one patient presented recurrence of the lesion, diagnosed due to an episode of thyroiditis 3 years after the endoscopic treatment. It was decided to repeat endoscopic treatment of the fistula, but the patient was lost from the follow-up.

Discussion

Branchial anomalies represent 20% of paediatric congenital malformations in the region of the head and neck, while those deriving from the third and fourth branchial arches are very rare (less than 1% of branchial anomalies). Although this complaint has been denominated PSF and/or remnant of the third or fourth branchial bursa, these names are confusing as they are rarely true fistulas, while some authors question their branchial origin. Due to these reasons some authors prefer to use the term pyriform fossa sinus tract.

Although it is present from birth, PSF mainly manifests in the first or second decade of life, and it is more common in males (at a ratio of 1.54:1).

There is clear predominance of the left side (93% vs 97% of cases) which is explained by the asymmetric development of the pharyngeal bursi during the embryonic stage, given that often the last right branchial body does not develop in the human species.

The most common form of presentation in children and adolescents is a recurring cervical abscess and suppurating acute thyroiditis, as was the case in our patients. Cervical infection is due to contamination of the internal orifice of the PSF. Clinically this presents as a cervical mass with signs of local inflammation, associated with odinophagia and fever. Thyroiditis is rare in children as the thyroid gland is highly resistant against infection (due to its high iodine content, rich vascularisation and fibrous capsule), so that PSF must be suspected in all children with thyroiditis.

As this disease is rare and its clinical presentation is non-specific, diagnosis is often delayed. In our cases the average interval between the appearance of the first signs and/or symptoms and diagnosis was 1.3 years. A high level of suspicion is extremely important for exact diagnosis and the correct treatment.

Imaging studies such as ultrasound scan and CT imaging are often performed to evaluate the cervical mass in a child, and they are very useful in determining the state of the thyroid gland, the site of the abscess, the extension of the lesion and, in the case of CT imaging, to visualise the pyriform sinus and fistula trajectory, using special manoeuvres to use air as a contrast medium (such as Valsalva’s manoeuvre). Nevertheless, secretion or tissue oedema may mask the fistula trajectory, hindering identification.

A barium oesophagogram may confirm the existence of PSF, with a sensitivity from 50% to 80%. It is recommended that it be performed at least 2 weeks after the cessation of
the inflammatory process, as in the acute phase it may give rise to false negatives.9

Optic fibre nasal endoscopy, carried out under local anaesthetic in the surgery, may sometimes reveal the opening of the fistula in the hypopharynx.6

The diagnostic technique of choice is endoscopic examination using a rigid instrument in the operating theatre and under general anaesthetic, as this makes it possible to confirm the diagnosis by means of direct visualisation of the tract orifice in the pyriform sinus.2,3,4,7,8 This has the additional benefit of permitting immediate and potentially definitive treatment.

In our experience the role of radiological examination was inconclusive, and the diagnosis of PSF was based on clinical suspicion and rigid endoscopy.

To prevent subsequent infections, complete resection of the fistula trajectory by cervicotomy, with or without hemithyroidectomy, has been recommended as the definitive treatment.6,8,11 Nevertheless, it may be hard to identify the fistula, and there is the risk of injuring adjacent structures such as the upper larynx and recurrent nerves.7

Several strategies have been described to aid identification of the tract during dissection. These include endoscopic channelling of the tract using a guide and the injection of pigment—such as methylene blue—through the fistula orifice.5,12 Even so, in some cases resection is incomplete and more than one intervention is required.7

Open surgery has a failure rate of 15% and a 6% risk of complications. The most common complications are vocal cord paralysis, a salivary fistula and infection.9

Different endoscopic techniques have been developed as an alternative to open surgery for the definitive treatment of these lesions. The most widely used of these is endoscopic electrocauterisation of the fistula tract opening, which was first described in 1998.6,7 Other techniques are laser, chemical cauterisation using trichloroacetic acid, chemical cauterisation using silver nitrate and fibrin.9,13,14

An endoscopic approach is less invasive and makes it possible to avoid the risks and cosmetic after-effects of open surgery. Complications are rare and recurrence may be treated using another cauterisation, giving good results over the long term.2,3,5-8 Another advantage of endoscopic treatment is that it shortens the stay in hospital, as it can be performed with a very short admission or even as an out-patient, thereby also reducing the economic cost.7

Depending on the technique used and the number of treatments, the success rates documented in the literature range from 78.9% to 100%.9 There is no evidence that any single technique is better than another.7 The only documented complication was temporary vocal cord immobility in 2 cases treated using chemical cauterisation by trichloroacetic acid.8,10 There were no reports of malignant conversion of the remaining tract following its obliteration.3

Endoscopic management may be considered to the first line treatment for PSF.5,9 As its morbidity is very low, several endoscopic attempts at obliteration may be made if symptoms recur. Up to 3 endoscopic attempts have been described before complete closure was achieved of the internal orifice of the fistula tract.3 Surgery is recommended in case of repeated failure of conservative treatment.3,8

Although our number of cases is low, the result is encouraging. The success rate was 85.7% and no complications arose. Endoscopic cauterisation with a radiofrequency electric scalpel was found to be a safe and effective procedure, and it may be considered as an option in the treatment of PSF.

Patients should be followed-up over the long term due to the possibility of PSF recurring.1 Patients must be advised to pay attention to recurring symptoms even years after an apparently successful procedure.

Conflict of Interests

The authors have no conflict of interests to declare.

References
